Human Monstrosities

Barton Cooke Hirst

George A. Piersol

Part III.

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HUMAN

MONSTROSITIES.

BY

BARTON COOKE HIRST, M.D.,

PROFESSOR OF OBSTETRICS IN THE UNIVERSITY OF PENNSYLVANIA,

AND

GEORGE A. PIERSOL, M.D.,

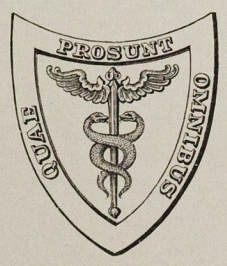
PROFESSOR OF HISTOLOGY AND EMBRYOLOGY IN THE UNIVERSITY OF PENNSYLVANIA.

PART III.

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AND

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CONTENTS.

	PAGE
CYCLOCEPHALUS	113
Ethmocephalus	113
Cebocephalus	113
Rhinocephalus	113
Cyclocephalus	114
Stomocephalus	114
Degrees of	115
The Eye	118
The Proboscis	120
Etiology	121
OTOCEPHALUS	126
Classification	127
OMPHALOSITIC MONSTERS	128
Etiology	129
Paracephalus	133
Acephalus	137
Asomata	139
Fœtus Anideus	139
COMPOSITE MONSTERS	143
Diprosopus	144
Diophthalmus; Distomus	145
Triophthalmus	147
Tetrophthalmus	148
Triotus	148
Tetrotus	149

PART III.

LIST OF PLATES.

PLATE XXI. Rhinocephalus.
" XXII. Cyclocephalus.
" XXIII. Skeleton of Cyclocephalus.
" XXIV. Paracephalus.
" XXV. Acephalus.
" XXVI. Acephalus.
" XXVII. Acephalus.
" XXVIII. Diprosopus.
" XXIX. Diprosopus. (Rear view.)

LIST OF ILLUSTRATIONS.

FIG.		PAGE
44.	Cyclocephalus	114
45.	The First Grade of Cyclocephalus	115
46.	The Second Grade of Cyclocephalus	115
47.	The Third Grade of Cyclocephalus	116
48.	Well-marked Cyclops	117
49.	Cyclops showing Single Eye with Fused Corneæ	117
50.	Cyclops with ill-formed Eye Socket and No Eye	118
51.	Diagrammatic representation of a Cyclops in Profile	119
52.	Formation of Anterior Medullary Groove	122
53.	Formation of Cephalic Vesicles	122
54.	Development of Cyclopean Brain	123
55.	Shape of Buccal Orifice in Cyclocephalus	124
56.	Otocephalus	126
57.	Implantation of the Second Allantois on the First	129
58.	Acephalus	134
59.	The Same, dissected	134
60.	Acephalus Monobrachius	135
61.	Anastomosis of Umbilical Vessels	136
62.	Peracephalus	137
63.	Mylacephalus	137
64.	Acardiacus Acormus	138
65, 66.	Acardiaci Acormi	138
67.	Fœtus Amorphus	140
68.	The Same, laid open	141
69.	The Common Placenta of Autosite and Omphalosite	142
70.	Diprosopus Asymmetros	144
71.	Diprosopus Distomus	146
72.	Diprosopus Triophthalmus	147
73, 74, 75.	Diprosopus Tetrophthalmus	148
76.	Diprosopus Triotus	148
77.	Diprosopus Tetrotus	149

PART III.

DESCRIPTION OF MALFORMATIONS

(*Continued*).

DESCRIPTION OF MALFORMATIONS.

MONSTROSITIES.

SINGLE MONSTERS — *Continued.*

Cyclocephalus (*Cyclops; Synopsia*). — Cyclocephalic or cyclopean monsters are thus described by Geoffroy Saint-Hilaire: "In the absence of the olfactory organs, more or less completely atrophied, the visual organs on both sides, imperfectly developed, sometimes altogether rudimentary, approach the middle line and almost always become intimately united with one another." Specimens of cyclocephalus are divided into five classes: ethmocephalus, cebocephalus, rhinocephalus, cyclocephalus, stomocephalus.

1. *Ethmocephalus.*[1]—In ethmocephalus the olfactory organs are not completely atrophied. There is a representative of the nose in the shape of a proboscis terminating anteriorly in two imperfect nostrils or in a single opening. There are two eyes and two eye-sockets.

2. *Cebocephalus.*[2]—In cebocephalus there is no nose, nor a representative of it. The interocular region is narrow and perfectly flat, but there are, nevertheless, two orbital cavities and two eyes.

3. *Rhinocephalus.*—In rhinocephalus the nose is represented by a proboscis inserted at the base of the forehead. Beneath

[1] ῥῑθμὸς, signifying here, root of the nose, and κεφαλὴ, head.

[2] κῆβος, monkey, and κεφαλὴ.

this proboscis is a single orbital cavity containing two orbits more or less intimately fused. (Plate XXI.)

4. *Cyclocephalus.*—In cyclocephalus proper there is a fusion of the orbital cavities and of the eyes without a vestige of a nose or of a proboscis. (Plates XXII. and XXIII.)

5. *Stomocephalus.*[1]—In stomocephalus there is the same deformity as in rhinocephalus, or in cyclocephalus; but, unlike these, the lower portion of the face is affected. The upper and lower maxillary bones are very defective, but the normal amount

FIG. 44.



Cyclocephalus.

of skin is provided, so that the latter, being in excess of the needs of the lower face, hangs in folds, especially at the region of the mouth, resembling somewhat the proboscis of rhinocephalus.

Having presented definitions of the divisions of cyclocephalus made by Geoffroy Saint-Hilaire, under names still employed by some writers, especially the French, we shall ourselves disregard them, being guided in the study of cyclocephalus in the main by the simpler and more rational system employed by Ahlfeld. According to this author, there are regular grada-

[1] *στόμα*, mouth, and *κεφαλή*.

tions of the deformity, from a simple approach of the eyes to one another, a narrowing of the face, and ill-development of the nose, to an ill-developed single orbital cavity with no eye at all. The different grades of cyclocephalus may be described as follows:

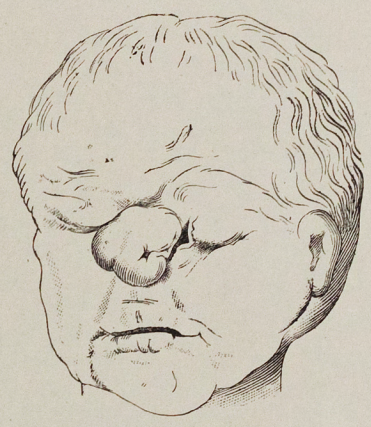
1. The eyes approach one another; the face is narrow, and the nose, while maintaining its normal situation, and, in part, appearance, is somewhat rudimentary. In a case described by Hecker and Buhl, the skull was very small, the bones were

FIG. 45.



The first grade of cyclocephalus.
(HECKER and BUHL.)

FIG. 46.



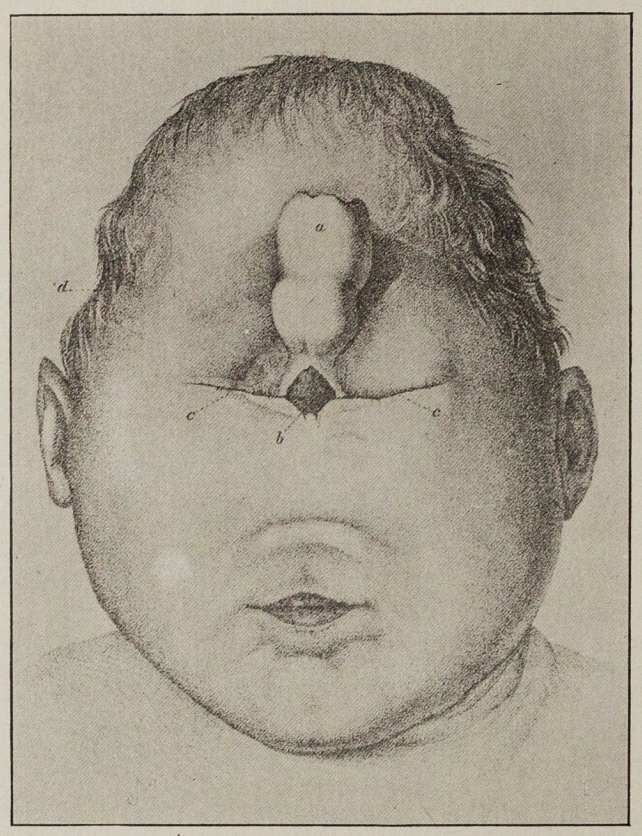
The second grade of cyclocephalus.
(AHLFELD.)

close together, and there was synostosis of the frontal suture, a condition almost always present in the most pronounced types of cyclocephalus. A cleft in the orbital plates opened into the nasal and oral cavities. The hemispheres of the brain were not separated from one another. The lateral ventricles were united in one large cavity filled with a large amount of fluid. (Ahlfeld.)

2. There is a closer approach of the eyes to one another. There is a mere rudiment of nose, but it is still between the eyes.

3. There is a union of the orbital cavities at their inner edge; but the eyelids still extend across the face on both sides. The nose is represented by a cylindrical body above the

FIG. 47.



The third grade of cyclocephalus. (CRAIG.) *a*, proboscis; *b*, fused orbits; *c*, palpebral fissures.

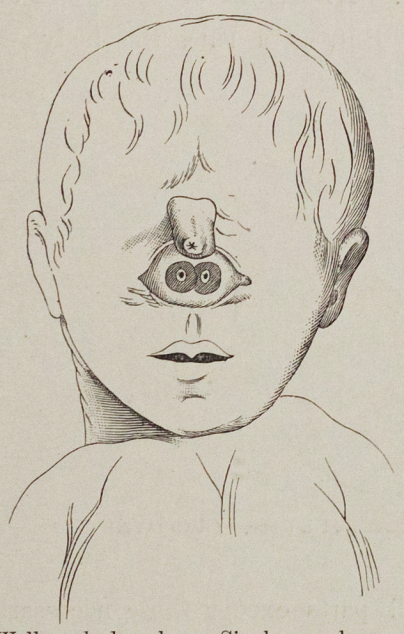
junction of the eye-sockets, composed of the skin structures of the nose, with possibly some cartilaginous formation at the base, or pedicle. (See Fig. 47.)

4. There is a complete union of the orbital cavities and a junction of the two orbits, the single eyeball, however, pos-

sessing two corneæ, two pupils, and two lenses. There may be an entire absence of proboscis, but it is usually present, always above the single eye.

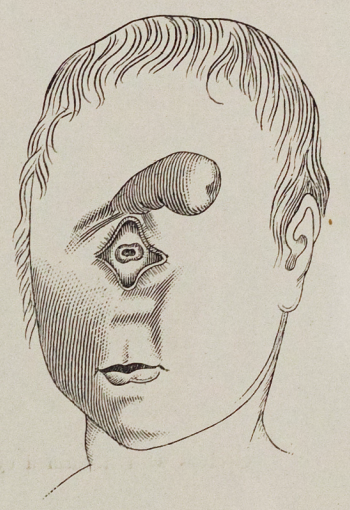
5. The corneæ of the single eyeball are united into a figure-of-eight space. The fused eyelids form a rhomboidal figure. There may still be two pupils and two lenses, or the latter may

FIG. 48.



Well-marked cyclops. Single eye shows duplex formation. (AHLFELD.)

FIG. 49.



Cyclops showing single eye with fused corneæ. (VROLIK.)

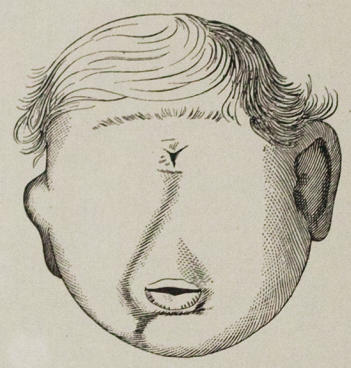
be single, but larger than common, and elongated in its transverse diameter. The single eye may be nearly or quite normal as far as external appearances go, and even an internal examination may show no duplication of any constituent parts, but there is commonly some indication of a duplex character, possibly a septum running antero-posteriorly, partly or wholly through the bulb, or perhaps an annular groove, running antero-posteriorly around the orbit. (Fig. 49.)

6. The cornea is not formed at all. The sclera covers the whole eyeball, and fills out the space between the two lids.

7. The eye-socket is very ill-developed and the orbit rudimentary or altogether absent (anophthalmia cyclopica).

8. The cyclops may show other deformities of the head, face, or distant regions of the body. Anencephalus is a rather frequently associated anomaly. The infant may exhibit also an otocephalic condition, or synotia. Anomalies of the trunk and extremities may be discovered. But, as a rule, the body will

FIG. 50.



Cyclops with ill-formed eye-socket and no eye. (RADDATZ.)

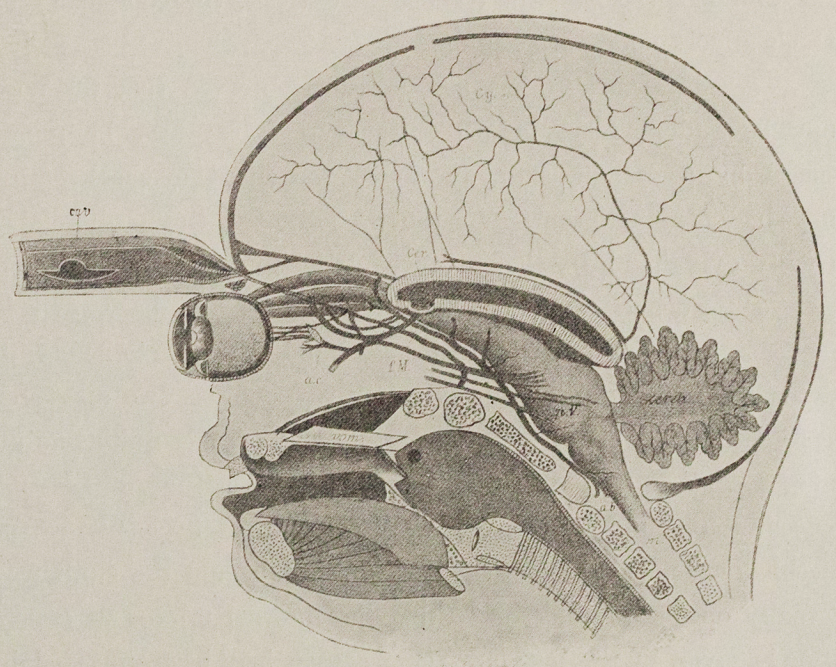
be apparently well formed in all parts except those necessarily involved in the cyclocephalic deformity.

One anomaly, however, polydactylism, is noted quite frequently; and, internally, anomalies of the heart and double uterus and vagina have been observed (Malherbe, Eustache, Craig, Debierre). Galloupe has reported an interesting case of cyclocephalus and hydrencephalocele anterior. The eye was on the anterior, lower part of the tumor, which involved the forehead and all the face from a point 0.5 cm. above the mouth.

The Eye.—In describing the different grades of cyclocephalus, based on the degree of approximation or fusion of the

eyes, mention has necessarily been made of some of the typical changes in the orbit and orbital cavity. There are other interesting conditions, however, to be noted in a closer study of the eye in cyclocephalus. The optic nerve is usually a single large trunk running to the orbital cavity. After entering the eye-

FIG. 51.



Diagrammatic representation of a cyclops in profile. *cer*, brain substance; *f M*, foramen of Monroe; *cy*, large space filled with fluid; *cereb*, cerebellum; *p V*, pons Varolii; *m*, medulla oblongata; *a b*, arteria basilaris; *a c*, arteria centralis retinæ; *vom*, vomer; *cav*, space lined with mucous membrane. (DELLA CHIAIE, from AHLFELD.)

socket the nerve trunk may divide into two branches, as in Craig's case. There may be no optic nerve at all, as in Renner's case, in which the eyes were also absent. The lenses and pupils may be double, although the eyes are fused into one globular mass covered with a single sclerotic and showing united corneæ. The Meibomian glands are usually to be seen

in the mucous membrane of the lids, which are commonly provided with eyelashes. If the eyes and eyelids are fused, a lachrymal caruncle may be found at the inferior angle of the rhomboidal figure made by the joined lids.

The orbital muscles have been described in one case (Eustache) as a "veritable plexus of muscular connections between the orbit and the walls of the orbital cavities, the complexity of which almost defies description."

The Proboscis.—It has been already observed that the nose may be rudimentary, but in normal position; may be represented by a cylindrical body above the fused eye-sockets, or may be entirely absent. In the second case, the proboscis is usually cylindrical in shape, is composed of skin, fat, and connective tissue, is somewhat pedunculated at the base, and is about 2 cm. long. It contains a blind pouch, clothed with mucous membrane, opening externally by one or two apertures representing the nares. The general appearance is quite like an infantile penis. In Palfyn's supplement to his edition of Licetus, indeed, the snout is labelled the male organ in a picture of dicephalus, one of the heads being cyclopean, and the skull was actually opened after death to find the internal organs of generation! Even in the last few years a case has been reported in which the writer refers to the penis-like body projecting from the forehead.

Many deviations from the description just given may be observed in individual specimens. Craig describes a cyclops in which the proboscis contained a single canal that, near its base, communicated with two rudimentary nasal cavities. These cavities were surrounded, except in front and above, by a bony capsule. The roof was formed by a thin bar of cartilage, and the dura mater. There was a bony median septum, reaching to the floor of the cavity, but not uniting with it. The outer wall

of each cavity contained some irregular spaces which might be regarded as rudimentary ethmoidal sinuses. Debierre describes a proboscis 2.5 cm. long and 4 cm. in circumference. On the free extremity was a depression at the bottom of which were two little openings communicating with the interior. In some cases the proboscis is very small and not perforated at all (Phisalix). It may be joined to the forehead by a very slender pedicle.

Etiology.—In describing the mode of formation of cyclocephalus, we shall present mainly the views of Dareste, recently published.[1]

Two opposite theories have been advanced to account for cyclocephalus; one, that there is a fusion of two eyes originally distinct (Is. Geoffroy Saint-Hilaire and Meckel); the other, that synopsia is an arrest of development, because normally there is at first the material for a single eye which later is differentiated into two (Huschke).

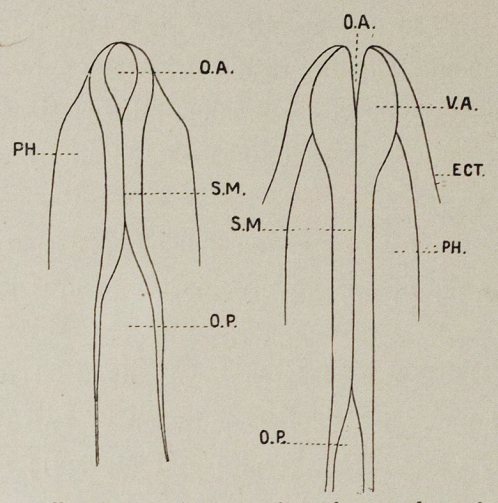
To understand the anomaly of development which results in cyclocephalus, it is necessary first to glance at the normal development of the fore-brain and of the eyes.

The medullary groove on the back surface of the embryo soon becomes a closed canal opened posteriorly and anteriorly. At the latter situation, three constrictions are developed, and between them the cerebro-spinal tract exhibits enlargements which become the three cerebral vesicles. The first, or anterior, enlarges laterally, so that the transverse becomes twice as great as the antero-posterior diameter. Constrictions then appear on both sides, ultimately forming two small lateral vesicles attached to the parent vesicle by pedicles. These are the primitive optic vesicles. At this time the small opening which has existed at

[1] "Mode de formation de la Cyclopie," Ann. d'Oculistique, 1891, pp. 171–182.

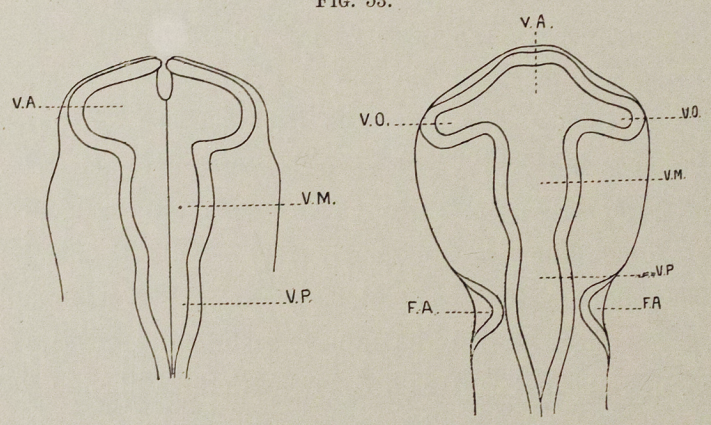
the anterior extremity of the parent vesicle, closes permanently. The optic vesicles pushing out laterally to the ectoderm are met

FIG. 52.



The anterior medullary groove in its earliest form, and at the period when the anterior vesicle is just beginning to develop. O.A. Anterior opening of the medullary groove. O.P. Posterior opening. S.M. Median junction of the wall. ECT. Ectoderm. PH. Pharynx. V.A. Anterior vesicle. (DARESTE.)

FIG. 53.

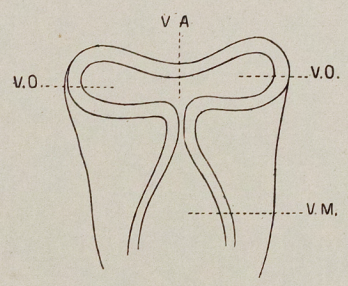


Diagrammatic representation of the formation of the cephalic vesicles. The three anterior vesicles are differentiated. V.A. Anterior vesicle. V.M. Middle vesicle. V.P. Posterior vesicle. V.O. Optic vesicles. F.A. Auditory fossæ. (DARESTE.)

by an invagination of the latter carrying a mass of cells that becomes detached and forms the crystalline lens.

The enlargement of the anterior cerebral vesicle has been accomplished, prior to the closure of the anterior opening, partly by the invagination of cells from the ectoderm which enter into the composition of its walls. After the closure of the anterior opening, the growth of the vesicle depends upon the multiplication of the cells contained in its walls. The movement of the ectodermic cells is a continuous one until the closure of the opening is effected, and each successive instalment of cells has perhaps the function of supplying some parts of those organs and tissues formed from the anterior vesicle. If, therefore, there

FIG. 54.



Development of cyclopean brain. V.A. Anterior vesicle. V.O. Optic vesicle. V.M. Middle vesicle.

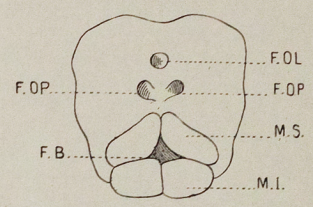
should be a premature closure of the opening the vesicle would be robbed of the cells necessary to its full development, and to the outgrowth later of organs derived from it. The ectodermic cells have an important rôle in the formation of the retinæ. At first they lie near to one another at each side of the anterior opening, but, pushed to either side by fresh relays of ingrowing cells, they come eventually to occupy the extreme lateral regions, in which are formed the optic vesicles, and after the anterior opening is closed are separated by tissues which will give birth to the cerebral hemispheres.

Dareste, from the study of young embryos affected with

cyclocephalus, declares that the first step in the deformity is the premature closure of the orifice of the anterior cerebral vesicle. If the closure is effected very early, but a single optic vesicle is developed in the median anterior region of the cerebral vesicle, and the degree of differentiation of the two orbits depends entirely upon the time at which the orifice is closed.

The ectoderm situated in front of the single orbit forms by invagination the single lens, or two, partially or wholly divided according to the extent of the division of the optic

FIG. 55.



Diagrammatic representation of the shape of the buccal orifice in cyclocephalus. F.OL. Olfactory fossa. F.OP. Optic fossæ. F.B. Buccal orifice. M.S. Superior maxillary. M.I. Inferior maxillary. (DARESTE.)

vesicles. Or, possibly, the lens formation is entirely in abeyance, and the eye is represented by a small mass of black pigment derived from the choroid.

As a necessary consequence of the anomalous development of the optic vesicle, there are anomalies of development in the face.

From the median situation of the eye in cyclocephalus, the fronto-nasal process cannot descend and occupy its normal position between the two branches of the superior maxillary. It remains above the single eye, and is more or less modified in development. The two divisions of the superior maxillary continue their growth until they meet one another in the median

line under the eye, without the interposition of the fronto-nasal process. This gives to the buccal orifice in the embryo, instead of the normal quadrilateral figure (see Fig. 1, Part I.), the shape of a triangle, with the base below, a shape characteristic of the cyclopean embryo. There is also an absence of those structures within the mouth which depend for their existence upon the descent of the fronto-nasal process. The fronto-nasal process, retained above the eye, shows itself peculiarities of development. The olfactory fossæ, instead of being separate, are fused in the middle line into a single cavity above the eye, without communication with the buccal cavity. Around this fossa there is commonly a development of ectodermal cells to form the proboscis, which is often perforated at its extremity. In case there is no such formation, the nose is represented only by a single rudimentary fossa above the eye.

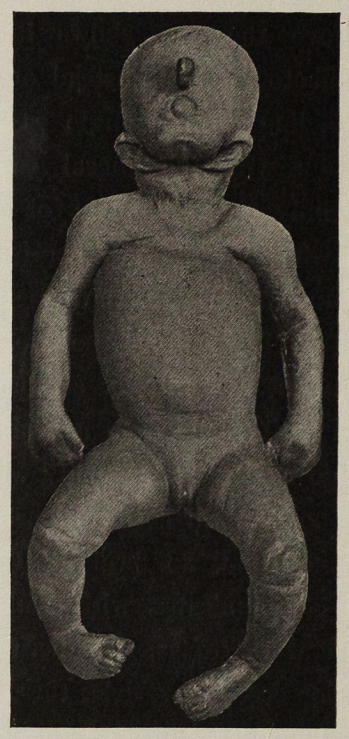
Within the cranium, the anterior cerebral vesicle shows an arrest of development. It remains in a vesicular condition, and presents a single large cavity filled usually with a large quantity of fluid. There are no cerebral convolutions, there is no corpus callosum, no corpus striatum, no fornix. The single cavity of the anterior vesicle communicates with that of the mid-brain, and there are no lateral ventricles. If there is a small amount of fluid in the anterior cerebral vesicle, there must be a marked narrowing of the bi-temporal diameter, but usually the forehead has an approximately normal width from the large quantity of fluid distending the walls of the vesicle.

According to the foregoing account, cyclocephalus is the result of an arrest in the development of the anterior cerebral vesicle. Dareste is disposed to believe that this arrest in development may often be due to anomalies in the development of the amnion, by which the latter exercises pressure or traction upon the anterior cerebral vesicle; especially is this

likely to have been the case when there are associated deformities in other parts of the body, like spina bifida, exencephalus, etc.

Otocephalus (*Agnathia; Synotia*).—In otocephalus, the lower jaw is altogether wanting or rudimentary. The ears approach one another, touch at their lower parts, and are even

Fig. 56.



Photograph of a cast in the Mütter Museum of the College of Physicians of Philadelphia.

united. The mouth is ill-developed, and may be represented by a minute orifice; in Underhill's case it was only large enough to admit a small probe; in the specimen possessed by the Mütter Museum of the College of Physicians of Philadelphia, and described by Smith, the mouth measured three millimetres in diameter. The upper jaw and the sphenoid may be also very

ill-developed. The hyoid bone is in some cases absent, in others perfect. The tongue is rudimentary or absent. The approaches to the trachea and œsophagus are sometimes open, sometimes closed. In Smith's case, the two external auditory canals communicated directly with the pharynx, the external auditory meati being represented by two slit-like openings 23 mm. apart, inclining downward and inward; there were no tympanic membranes.

Otocephalus is very rare in the human species, but seems to be not infrequent among sheep. The agnathia is usually joined with synopsia and often with hemicrania. The fœtus cannot live after birth on account of the impossibility of ingesting food, and often of respiring.

According to the classification of Geoffroy Saint-Hilaire, the following subdivisions of otocephalus may be made:

1. *Sphenocephalus.*—The two eyes are well separated; the ears are united under the head; the jaws and the mouth are distinct; the sphenoid is altered in shape, so that it is analogous in form to what is found normally in birds. Hence the name of this variety.

2. *Otocephalus.*—There is some degree of synopsia; the ears are nearly or quite joined together; the maxillæ and mouth are distinct; there is no proboscis.

3. *Edocephalus.*[1]—There is synopsia; also a proboscis; beneath the eye is a small opening which might be taken for a mouth, but is the fused external auditory meati, in the median line; there is, consequently, no mouth and very rudimentary jaws.

4. *Opocephalus.*[2]—There is synopsia, with the characteristics of otocephalus, but there is neither mouth nor proboscis.

[1] *αἰδοῖον*, genitalia; from the resemblance of proboscis to penis.

[2] *ὤψ, ὠπὸς*, the eye.

5. *Triocephalus.*[1]—There is a most extensive arrest of development. There is no ocular, nasal, or buccal apparatus. The head is little more than a small, spheroidal mass, covered uniformly with skin. At the junction of the head with the neck is a small opening, the fused auditory meati, and, extending to either side, are the auricles.

The cause of otocephalus, or agnathia, is an arrest in the development of the lower processes (Meckel's cartilages) of the first branchial arches. The arrest of development may affect this portion, as well as any other part of the embryo, without the interposition of any mechanical interference with normal growth; but, according to Dareste, the primary cause may be an arrest of development in the cephalic fold of the amnion, whereby the cephalic extremity of the embryo may be subjected to a pressure which will prevent its perfect development.

Order II. Omphalositic Single Monsters.

The preceding anomalies of development in the fœtus and infant have been included under one general head—autosites; that is, embryos capable of an independent existence in the womb. The omphalosites are embryos or fœtuses dependent for their imperfect growth in the uterus, upon another embryo or fœtus, usually well developed, that supplies the circulation of blood for both by means of extensive and intimate anastomoses of the umbilical and placental vessels.

Two divergent views are entertained by modern teratologists as to the formation of omphalositic monsters: one, that of Claudius and Ahlfeld; the other, that of Breus.

[1] Etienne Geoffroy Saint-Hilaire called these specimens "triencephales." His son retains the term, modified as above.

The first is clearly enunciated by Ahlfeld. Two healthy embryos develop upon a single yolk. The allantois of one is formed a few hours sooner than that of the other. The allantois of the former has reached the internal surface of the primary chorion and grown entirely or partly around it, when the allantois of the latter endeavors also to reach the same destination. If the first has spread all over the inner surface of the chorion, the second can establish no direct connection with it, but must be inserted in the first-formed allantois (see Fig. 57). But if a

FIG. 57.



The implantation of the second allantois on the first. *A*, the first embryo; *B*, the second. (AHLFELD.)

small area of the chorion remains free, the second allantois can establish a connection with it, whether it corresponds with the decidua vera or reflexa. In the latter case, the second embryo does not acquire a distinct placenta, but must utilize the allantois of the first for the insertion of its vessels. In the second case—*i. e.*, if the small portion of unoccupied chorion corresponds with the decidua vera, the second embryo gains a small portion of the common placenta for the insertion of its vessels. Dependent upon this difference of arrangement in early embryonic life, Ahlfeld classifies the omphalosites into true, or pure, and secondary allantoic parasites.

In the first variety, the second allantois, as already described, implants itself upon the first. The connection between the bloodvessels of the two becomes intimate and extensive, but their relations depend upon the growth of the second amnion. If this is large and spreads over the surface of the placenta, the umbilical bloodvessels of the second embryo apparently enter the placenta directly from the cord; if it remains small, the bloodvessels of the cord must join those of the first embryo by a velamentous insertion.

In the second variety—secondary allantoic parasites—the second embryo implants its allantois partly in the allantois of the first, partly in the chorion. One of the primitive aortas in the second embryo then establishes a connection with the allantois of the first, while the other aorta communicates with the chorion. For a while the two circulations continue side by side, but soon the chorionic vessels atrophy and the second embryo derives its nutriment from the communication between its vessels and those in the allantois of the first embryo. In other words, the allantoic placenta of the second embryo grows at the expense of the chorionic. The latter undergoes atrophy and fatty degeneration. If the chorionic placenta of the second embryo develops and the allantoic withers, or is from the first quite subordinate, there is seen the ordinary common placenta of unioval twins, with capillary anastomoses, or the junction of larger vessels, perhaps, but not the formation of an omphalosite.

In consequence of the inosculation of the umbilical vessels of the omphalosite into the large umbilical and placental vessels of the autosite, the two blood-currents meet in opposing streams. The heart power of the first embryo, or autosite, being the greater, the blood-current of the second is reversed, flowing back through the umbilical artery to the trunk and heart, passing through the latter as though it were a simple

continuation of the blood-channel. Consequently the heart atrophies or finally disappears, becoming simply a passive part of the bloodvessels running through it. Following the failure of cardiac development, there is absence or imperfect development of other internal organs and of important regions of the body, as the head and upper extremities.

Breus has, in recent years, attacked the theory of Claudius and Ahlfeld as untenable. In place of it, he offers the following explanation:

"The acardiaci, on account of early disturbance of their embryonic development, are such highly defective products of conception that they are not capable of an independent circulation or of an intra-uterine nutrition; but they, nevertheless, continue to develop and to grow, because a supplementary circulation maintains their tissues in a living and growing state. They are usually found in twin ova along with a normal fœtus, for under these circumstances, by anastomosis of the umbilical vessels of both fœtuses, the vicarious circulation is most easily established. But it is possible, in rare cases, of course, for the vicarious circulation to be established in a more complicated manner."

According to Breus, therefore, the omphalosite dies and no longer possesses an independent circulation. Then the blood-current of the autosite enters the vessels of the omphalosite and keeps it nourished to an imperfect degree. According to Breus, omphalositic monsters should be very common, if their production depended upon the reversal of a weak blood-current by a stronger, for in all unioval twins anastomoses between the vessels of the two cords are invariably demonstrable and one fœtus is usually better developed than the other.

From the absence of the heart in many omphalosites the name acardiacus is often employed, especially by German writers,

to designate the condition. Ahlfeld, however, collected fourteen cases in which the heart was present in some degree of development, and in some of the cases, at least, was functionally active until birth, as proved by the presence of trabeculæ carneæ. We think, therefore, that omphalosite, the name employed by Geoffroy Saint-Hilaire, is more appropriate than acardiacus, and should be retained.

There are certain features common to all forms of omphalosites that we shall notice before proceeding to a detailed description of the different forms. From the small size of the efferent vessel of the omphalosite, and from the deficient *vis a tergo* in the venous circulation, due to the great distance the blood must be propelled by the autosite's heart, there is always a more or less sluggish circulation in the parasitic twin. Consequently, there is an overgrowth of connective tissue, especially the subcutaneous, and in this situation there are developed lacunæ and cysts. For the same reason, œdema of the omphalosite is almost constantly noted. Possibly from the same cause a very advanced fatty degeneration of the muscular tissue has been observed in one case by A. R. Simpson.

In the sac of the parasite there may be oligohydramnios, while polyhydramnios is found in the sac of the autosite. But the reverse was seen in the case of Pasquali and Bompiani.

The parasite is usually born last, with the placenta, but exceptions to this rule are noted by Ross and Slyman. In their cases the autosite was delivered last.

There is scarcely ever any difficulty in the delivery of the parasite, which is almost always undersized, and may be a mass of flesh no bigger than the clinched fist. But in Ross's case, the œdema of the parasite obstructed delivery so seriously that it was necessary to amputate a thigh, seize the pelvis with a cranioclast, and use great force to extract the body.

The autosite also sometimes presents anomalies of development. In a specimen recently presented to our collection by Dr. John Palmer, there was a cleft palate and hare-lip, and, strangely enough, the omphalosite was much the larger fœtus of the two, the weights being respectively 2 pounds and $1\frac{1}{4}$ pounds.

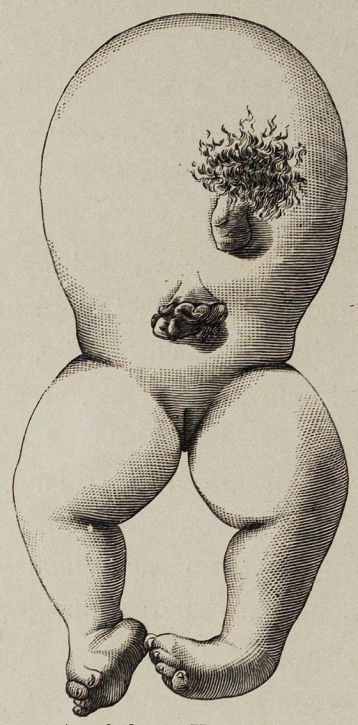
One might, however, expect a retardation of growth in the autosite, for its blood must be vitiated by the return current from the omphalosite emptying directly into its umbilical vein.

According to Geoffroy Saint-Hilaire, omphalositic monstrosities are divided into paracephalic, acephalic, and anideous specimens. To these must be added a fourth, asomatic, which was not recognized by Geoffroy Saint-Hilaire.

Paracephalus (*Acardiacus Anceps*).—In paracephalus is seen the highest development of which an omphalosite is capable. There are four extremities, always, however, more or less imperfect, especially in the digits. There is a head, but the cranium and face are imperfectly formed. There are cervical vertebræ, but they may be so rudimentary that the head is not distinctly separated from the trunk. The diaphragm is often defective; the lungs are absent or rudimentary. The assertion of Ahlfeld, that the heart, though rudimentary, is always present, is inaccurate, for in well-developed paracephali described by Koch, Doran, and Breus, there was no trace of a heart. The genito-urinary organs show usually the least abnormality. The cerebrum shows defective development; the cord and medulla are not usually affected. The paracephalus is one of unioval twins; the sex of both, therefore, is always the same and is usually feminine. According to Tarnier and Budin, the twins are always born prematurely. The paracephalus has never shown a sign of life after birth. (Plate XXIV.)

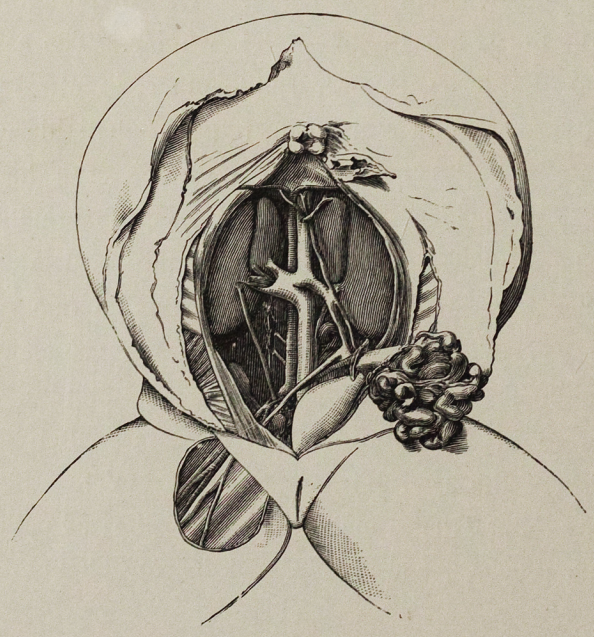
Geoffroy Saint-Hilaire makes three divisions of paracephalus: paracephalus proper, omacephalus, and hemiacephalus.

FIG. 58.



Acephalus. (VEENDAM.)

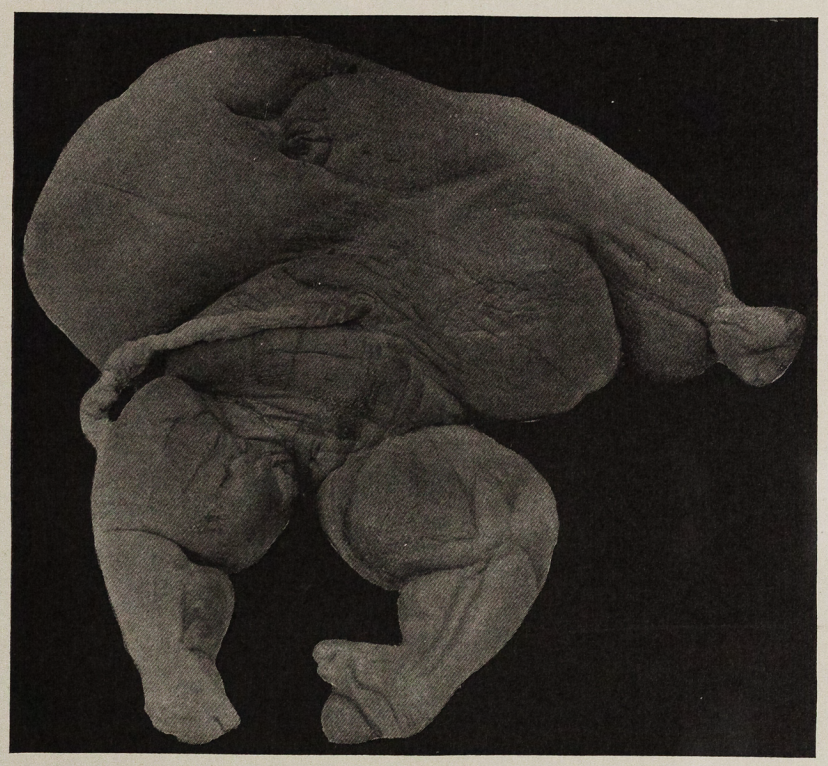
FIG. 59.



The same, dissected.

1. *Paracephalus* is characterized by a head very imperfectly formed, more or less defective in all its parts, but exhibiting rudiments of the cranium and of the organs of sense, and possessing even a mouth and buccal cavity.

FIG. 60.



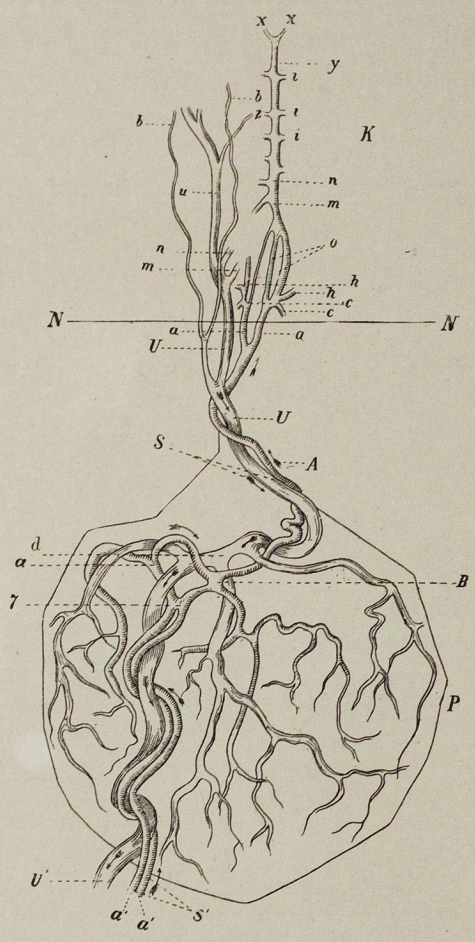
Acephalus monobrachius.[1] (PALMER.)

2. *Omacephalus* shows more imperfect development. The upper extremities are wanting. In other respects there are the characteristics of paracephalus.

3. *Hemiacephalus* is the lowest grade of development in paracephalus, closely approaching true acephalus.

[1] For a full description of this specimen and its dissection, see Hirst, "Two Recent Additions to the Teratological Collection in the Wistar and Horner Museum." University Medical Magazine, July, 1892.

FIG. 61.



Showing anastomosis of umbilical vessels and the distribution of the blood-supply to the omphalosite. (FELSENREICH and HOLL.) *P.* Common placenta, or, rather, placenta of autosite. *S.* Cord of the acardiacus. *S'.* Cord of the autosite. *U' a' a'.* Vein and arteries of the latter. *A.* Artery, *U.* Vein, of the cord of the acardiac. *a* and *B.* Anastomosis of the umbilical artery of the acardiac with that of the autosite. 7. Anastomosis of both umbilical arteries. ⟶ Direction of the blood-current. *d.* Inosculation of the two umbilical veins. *NN.* Entrance of the umbilical vessels in the umbilical ring. *K.* Circulatory system of the acardiacus: *b b*, veins to the anterior thoracic wall; *u*, vein on the spinal column, continuation of *U*; *z*, vena hemiazygos; *n*, vena renalis; *m*, vena mesenterica; *a a*, art. umbilic.; *c c*, art. crurales; *h h*, art. hypogastricæ; *y*, aorta; *x x*, its terminal branches; *i i*, art. intercost.; *n m*, art. renalis et mesenterica; *o*, divergence and re-convergence of the right umbilical artery.

ACEPHALUS.—There is complete absence of the head and usually of the upper extremities. The latter may be represented by stumps covered by the skin of the body. Occasionally one upper extremity is present, though not perfect (acephalus monobrachius, Plate XXVI.). Acephalus is the commonest condition among the omphalosites.

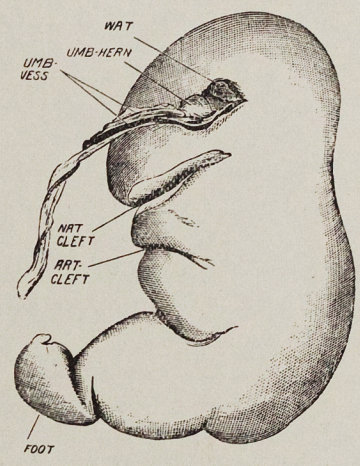
In acephalus there is often a rudimentary heart, usually with two chambers. The neighboring organs and structures,

FIG. 62.



Peracephalus. (VROLIK.)

FIG. 63.

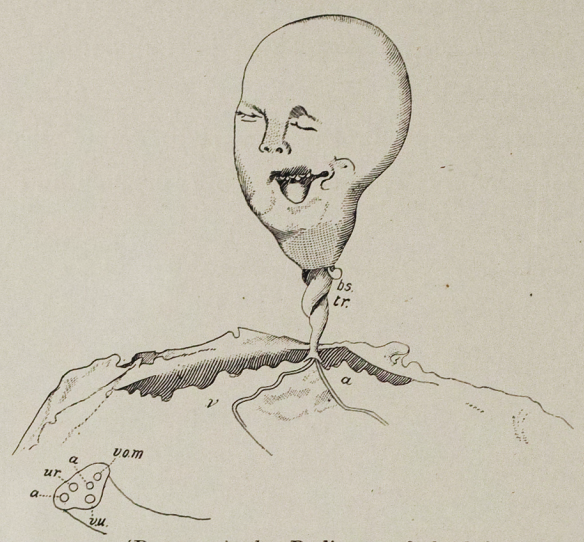


Mylacephalus. (DORAN.)

as the lungs, pericardium, trachea, diaphragm, sternum, parts of the ribs, vertebræ, etc., are very defective or altogether absent. The abdominal and pelvic organs and the lower extremities are best developed, because by the reversal of the circulation, the freshly aërated blood flows first to them through the two allantoic arteries. There is always a median fissure of the thorax. The genito-urinary apparatus is usually well developed (Plate XXV.), but in Simpson's case it was absent. The cord is inserted in the middle of the very short belly, and there is often a hernia into the umbilical cord. Sometimes above the

insertion of the umbilical cord there is a sac-like dilatation, which is often the remnant of an umbilical hernia, rarely the

FIG. 64.



Acardiacus acormus. (BARKOW.) *bs.* Rudiment of the left upper extremity. *tr.* Rudiment of intestine. *a a a.* Arteries. *v.* Vein. *v.u.* Umbilical vein. *v.o.m.* Omphalo-mesenteric vein. *ur.* Urachus.

FIG. 65.



FIG. 66.



Acardiaci acormi. (AHLFELD'S Atlas.)

remnant of an ectopic heart (Plate XXV.). There is usually only an afferent and an efferent vessel in the cord, but there

are two arteries internally, one of which supplies the lower extremities, the other, the upper part of the body, running through the rudimentary heart, if it exists.

Of acephalus, Geoffroy Saint-Hilaire makes three divisions:

1. *Acephalus.*—This is the highest form of development in the acephali. There is wanting the head, but there is at least one superior extremity, and the thorax is fairly well developed. There may be some cervical vertebræ and rudiments of the head hidden under the skin.

2. *Peracephalus.*—The upper part of the trunk is defective. There are no superior extremities. The body may be reduced to the pelvis and lower extremities.

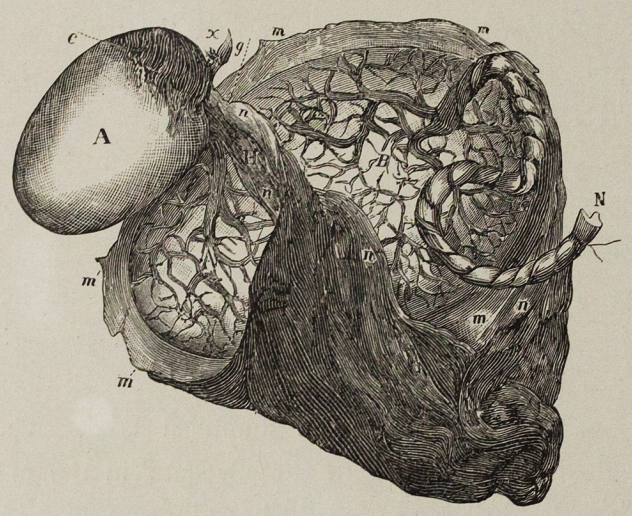
3. *Mylacephalus.*—There is such imperfect development of the fœtus that the condition is but a degree above that of fœtus amorphus or anideus.

Asomata (*Acardiacus Acormus*).—In the asomatic, acormic, or trunkless omphalosites there is a more or less distinct indication of a head, which is never, however, well formed. In none of these cases is there an umbilical cord. The vessels run directly from the head to the placenta in the membranes. Beneath the head is a sac in which rudiments of body organs may be found. Of all the omphalosites, this is by far the rarest. We have found but one reference to an acardiacus acormus in the *Index Medicus* for thirteen years. It is, indeed, one of the rarest of all monsters.

Fœtus Anideus (*Fœtus Amorphus*, Gurlt).—In this, the lowest form of omphalosite, the parasitic fœtus is reduced to a shapeless mass of flesh covered with skin. There may be a bunch of hair and a slight prominence to denote the cephalic end of the mass. Internally there is apt to be a more complicated structure than one would expect from the external appearance. Single bones, especially those of the pelvis and

some of the vertebræ, more or less well developed, may be found. There are portions of the gut, ending in blind pouches above and below. The subcutaneous connective tissue is usually hypertrophied, cystic, and œdematous. In Neugebauer's specimen, however, this was not the case. There is usually no trace of a heart (Ahlfeld says never), but in Neugebauer's typical case of amorphous fœtus, in which the parasite was a mass no

FIG. 67.



Fœtus amorphus attached to the placenta. (NEUGEBAUER.) *A*. Fœtus amorphus. *B*. Placenta with single chorion and double amnion. *mmmm*, *nnnn*. Membranes. *g*. Umbilical cord of the fœtus amorphus. *N*. Umbilical cord of the autosite. *c*. Hair on cephalic extremity of omphalosite. *H*. Fold of amnion.

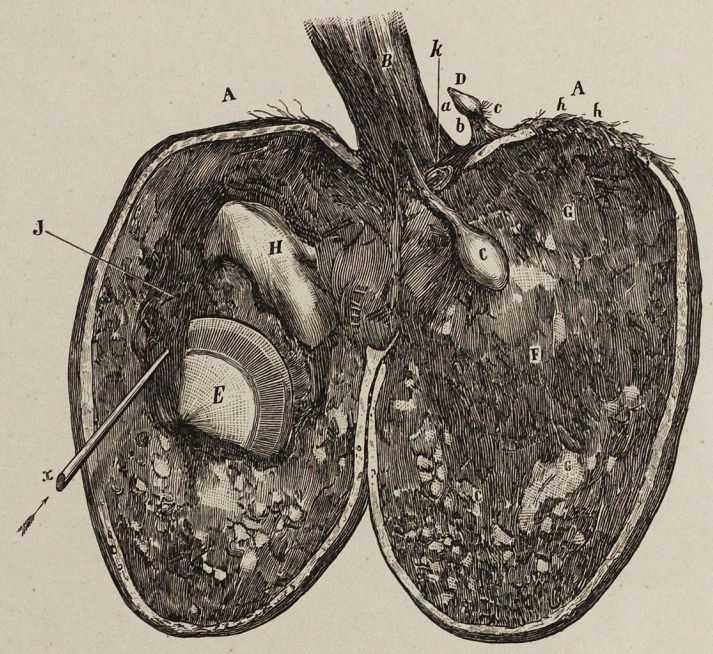
bigger than a clinched fist, attached to the placenta by a pedicle 2 cm. long, there was a single-chambered heart.

There are two small vessels in the cord, artery and vein. Often there is no cord, but the vessels of the parasite run in the membranes.

The Placenta, Cord, and Membranes of Omphalosites.—The placenta is always common to both fœtuses, the parasite and

autosite. Usually there is a common chorion, but in the case described by Pasquali and Bompiani there were two chorions. There are usually two amnionic cavities, sometimes a single one. Ahlfeld says that the parasite always has the lesser quantity of liquor amnii, but in the case just quoted the

FIG. 68.

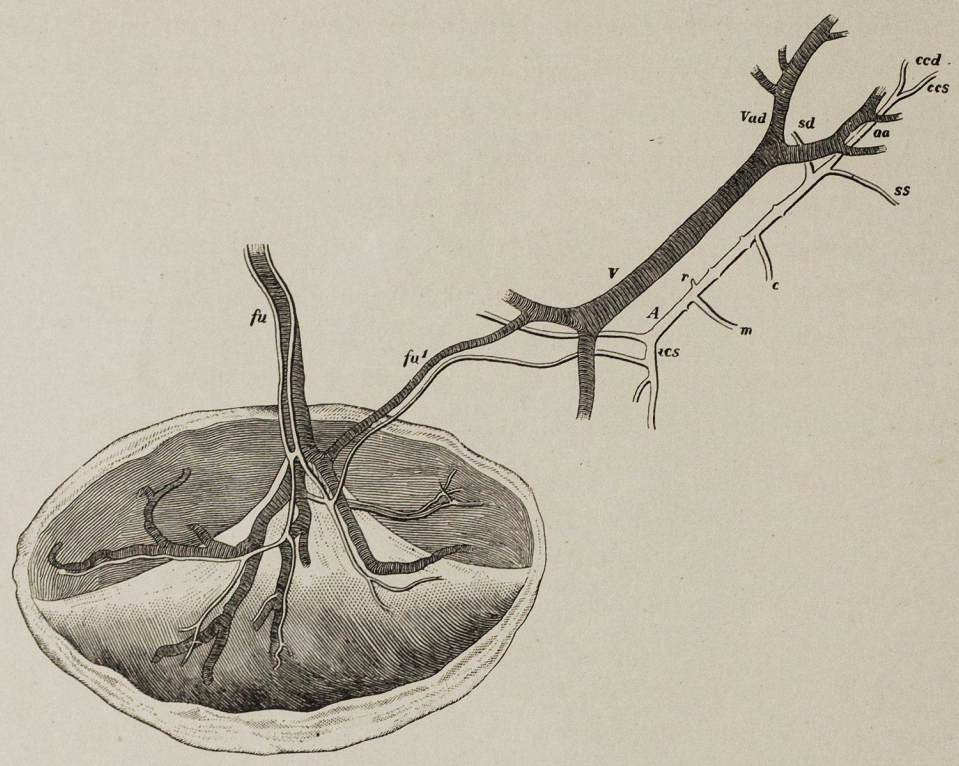


The same, laid open. *A A.* The two halves of the monster. *B.* The connection with the placenta. *C.* Rudimentary single-chambered heart. *D.* Penis-like rudiment of the head, with hairy growth (*b c*). *E.* Rudimentary iliac bone, with bony centre and cartilaginous periphery. *F.* Connective tissue. *G J.* Traces of muscle tissue. *H.* Rudiments of the lower lumbar vertebræ. *x.* Spinal canal with nerve tissue (a probe shows direction of canal). *h h.* Hair. *k.* Opening of vein.

amnionic cavity of the parasite was the larger. In this same case the umbilical vessels of the parasite ran free for 2 cm. from the placenta, and then in the membranes to the umbilical ring. Very often the umbilical vessels of the parasite have a velamentous insertion. In the junction of the umbilical vessels

from the two fœtuses it is the rule for the main artery in the umbilical cord of the autosite to run itself, or by a main

FIG. 69.



The common placenta of autosite and omphalosite, with a sketch of the blood-vessel system of the omphalosite in connection with the placenta. (BREUS.) *fu.* Cord of autosite. *fu'.* Cord of omphalosite (the veins are shaded, the arteries white); the umbilical vein of the omphalosite empties into the right common iliac vein, the umbilical artery into the left common iliac artery. Both run from the cord along the bladder to these vessels. *V.* Chief vein of the body, corresponding to the vena cava. *A.* Aorta. *ics.* Art. iliac. comm. sinistra. *m.* Art. mesariaca. *r.* Art. renalis. *c.* Art. cœliaca. *sd.* Art. subclavia dextra. *ss.* Art. subclavia sinistra. *aa.* Art. anonyma. *ccd.* Art. carotis comm. dextra. *ccs.* Art. carotis comm. sinistra. *Vad.* Vena anonyma dextra.

branch, directly to the umbilical artery of the parasite, sometimes running across the fœtal aspect of the placenta, sometimes

joining with the artery of the omphalosite by a Y-shaped junction of the cords before either reach the placenta. In Breus's case both cords had a velamentous insertion in their respective amnions, the latter by their junction making a fold at the middle of the placenta, in which the vessels of the two cords anastomosed some 3 cm. above the placental surface.

The vein of the parasite takes often a more tortuous course, uniting ultimately, however, directly with the umbilical vein of the parasite or with a large branch of it. Sometimes the connection between the veins is by means of numerous small branches of each. The cord of the omphalosite is usually short, sometimes extremely so. Ahlfeld has said that "the better developed the heart of the acardiacus, the more likely will its cord be inserted, by that of the autosite, in the centre of the placenta." Breus's case, however, in which there was no heart at all, shows the inaccuracy of this statement.

From the extra work thrown upon the heart of the autosite in the propulsion of blood through two bodies, it exhibits considerable hypertrophy, as was demonstrated by Meckel, who, with Hempel, in 1850, was the first to describe truly the nutrition and circulation of omphalosites. (Ahlfeld, p. 46 of the text.)

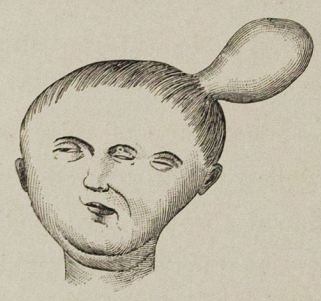
COMPOSITE MONSTERS.

By this term we designate all specimens showing some degree of duplicity in the main regions of the body. "Compound monsters" and "double monsters" are synonymous terms. Fisher uses the words diploteratography and diploteratology to denote the description and diagnosis of special forms of double monsters and the study or science of double monstrosities in general.

We shall not concern ourselves in this section with the genesis of composite monsters. For information on this point the reader is referred to the chapter on the Production of Malformations in Part I. We shall simply describe the different forms of double monsters, following, in our descriptions, the classification employed by Förster.

Diprosopus (*Iniodymus; Opodymus; Heteroprosopus*).—Diprosopus signifies double-faced. The duplicity of the face and head varies from a junction of two distinct heads and faces by their posterior and proximal lateral surfaces to a very slight

Fig. 70.



Diprosopus asymmetros (heteroprosopus). (Förster.)

trace of duplex formation externally. The double formation is confined to the head and does not affect the neck. The forms with completely separated faces are more common among domestic animals than in man. The partial separation of the faces is most common in calves and cats, less in man and among sheep, still rarer in swine, goats, and horses. It has never been observed in dogs. The diprosopic monster has been found in birds, fishes, and snakes, and has been seen in the tapeworm.[1]

[1] Fisher: Diploteratology. A work to which we are much indebted.

Of 500 monsters observed by Förster, only 29 were diprosopic—less than 6 per cent. Of the 29, 16 were females and 6 males; in 7 the sex was not stated. Of 5 cases examined by Fisher, 3 were females, 2 males. There is often hydrocephalus, acrania, defective development of the brain, and spina bifida. Mainly on this account the diprosopi are not viable. But infants have been described which lived fifteen minutes, several weeks, and, in one case, seven months, with a good prospect of continued existence (Regnault, quoted by Fisher). Although diprosopi are apparently normal in development below the neck, internally defects of development are common. In many cases a diaphragmatic hernia on the left side is noted. Anomalies of the heart and large bloodvessels have also been observed.

Usually the two faces are symmetrical; occasionally there is asymmetry. The relation of the facial axes is irregular: sometimes parallel, sometimes converging downward, again upward. The axial planes of the face meet behind at varying angles.

There appears to be no difficulty in the delivery of the head. In one case embryotomy was necessary in the extraction of the body by the feet, but the head was extracted without trouble. (Mays.)

The diprosopi have been classified as follows:

1. Diophthalmus.
2. Distomus.
3. Triophthalmus.
4. Tetrophthalmus.
5. Triotus.
6. Tetrotus.

1. *Diprosopus Diophthalmus;* 2. *Diprosopus Distomus.*—Fisher has described the first case of diprosopus diophthalmus, or

distomus, reported in English. There was anencephalus. The eyes were widely separated by what appeared to be a broad nose with two nares far apart. There were two triangular openings, one on each cheek, separated more than an inch by a broad chin, in the centre of which were two dimples.

FIG. 71.



Diprosopus distomus. (BUCH.)

On closer study, the nose was found to be duplex, with two imperforate nares adjoining in the middle line. The two triangular openings were mouths, communicating with a common oral cavity, in which there were two tongues joined at an angle of 80° at their posterior thirds. There was a single epiglottis, trachea, larynx, and œsophagus. The upper maxilla was apparently single, though the palate was very broad. The

lower maxilla was double, the inner rami were joined but not fused.

Occasionally the external evidences of duplicity are slighter than in the case just described. Fisher refers to Gurlt's specimen of a lamb in which there was a double tongue and an indication of duplication in the lower jaw, but nothing more. On dissection, however, there were discovered double formations in the bones at the base of the skull, an additional set of corpora quadrigemina, a double pineal gland and pituitary body.

FIG. 72.



Diprosopus triophthalmus. (SÖMMERRING.)

3. *Diprosopus Triophthalmus.*—In this variety there are three eyes, the median globe showing distinct evidence of duplicity in its long transverse diameter, double iris, cornea, and pupil. The sclerotic coat is apparently single, but internally, as in many cyclocephali, there is a sclerotic septum, the result of the imperfectly differentiated inner sclerotic walls of the orbits. The eyelids, though usually single above and below, show also, on close examination, signs of duplex formation. There are often other deformities of the face and head, such as acrania, and, as in MacDonald's case, hare-lip and cleft palate in the two mouths.

4. *Diprosopus Tetrophthalmus.*—In this variety the differentiation of the two faces is further advanced, but there are still but two ears. These specimens are quite different in appearance, as the facial axes are parallel, converge below or converge above.

FIG. 73. FIG. 74. FIG. 75.



Diprosopus tetrophthalmus. (FISHER, SÖMMERRING, FISHER.)

5. *Diprosopus Triotus.*—In this variety there are usually four eyes and always three ears; the median auricle, distorted in size and shape, consisting of the two adjoining ears of the double face, fused into one. In Genersich's case the median

FIG. 76.



Diprosopus triotus. (SÖMMERRING.)

ear was represented by a papillary projection, the size of a bean, the pedicle of which sprang from a fold between the two faces. In this same case, the four nostrils and two mouths opened into a single pharynx. The œsophagus and larynx were single.

6. *Diprosopus Tetrotus.*—In this variety, the intermediate stage between diprosopus and dicephalus, there are four eyes, and, as the name indicates, four ears. (Plates XXVIII. and XXIX.[1]) There is found in these specimens considerable duplicity in the cervical region. There are sometimes two

FIG. 77.



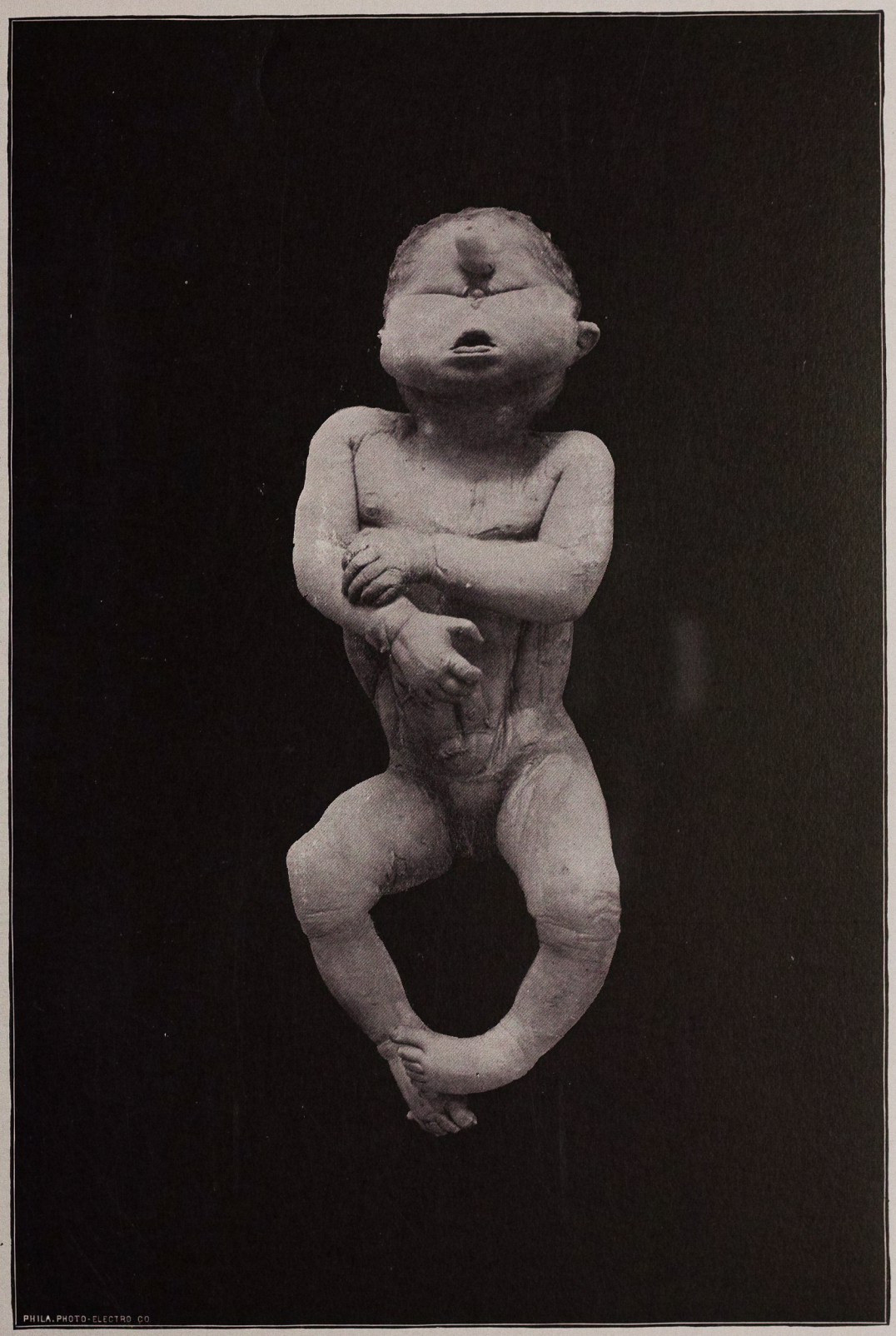
Diprosopus tetrotus. (FLEMING.)

pharynges and two œsophagi, the latter uniting, however, before entering the single stomach. The larynx and upper part of the trachea may also be doubled. Usually, however, there is a common pharynx, and the structures immediately

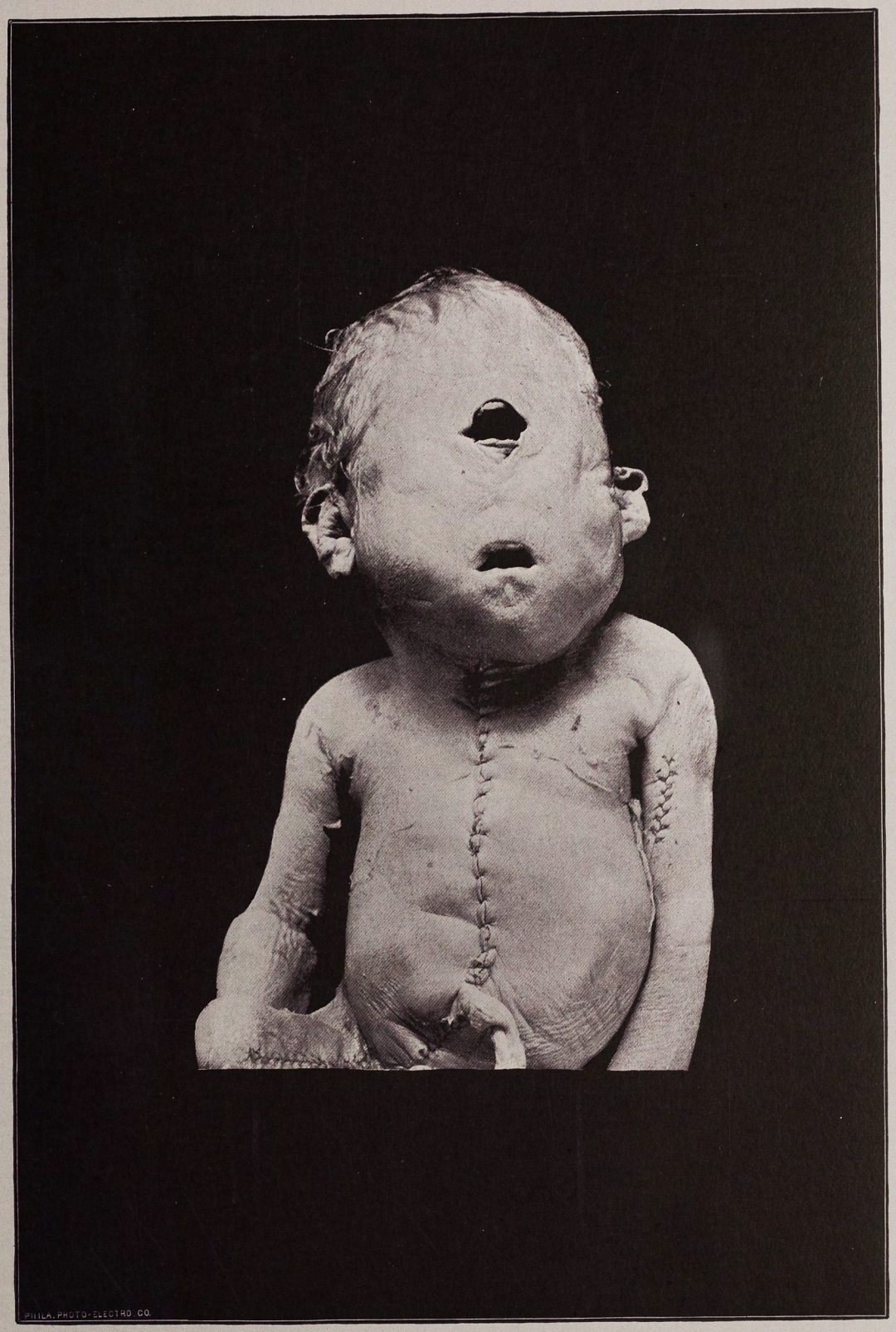
[1] The specimen represented in these plates has four ears; the two adjacent are smaller than the outer ears, but are distinct and separate.

below are single. Diprosopus tetrotus is extremely rare in the human race. Fisher says he knows of but a single specimen. We see no other mentioned by Ahlfeld, up to 1880, and in the *Index Medicus* for the past thirteen years we found a reference to only one other case, that of Fleming.

PLATE XXI.



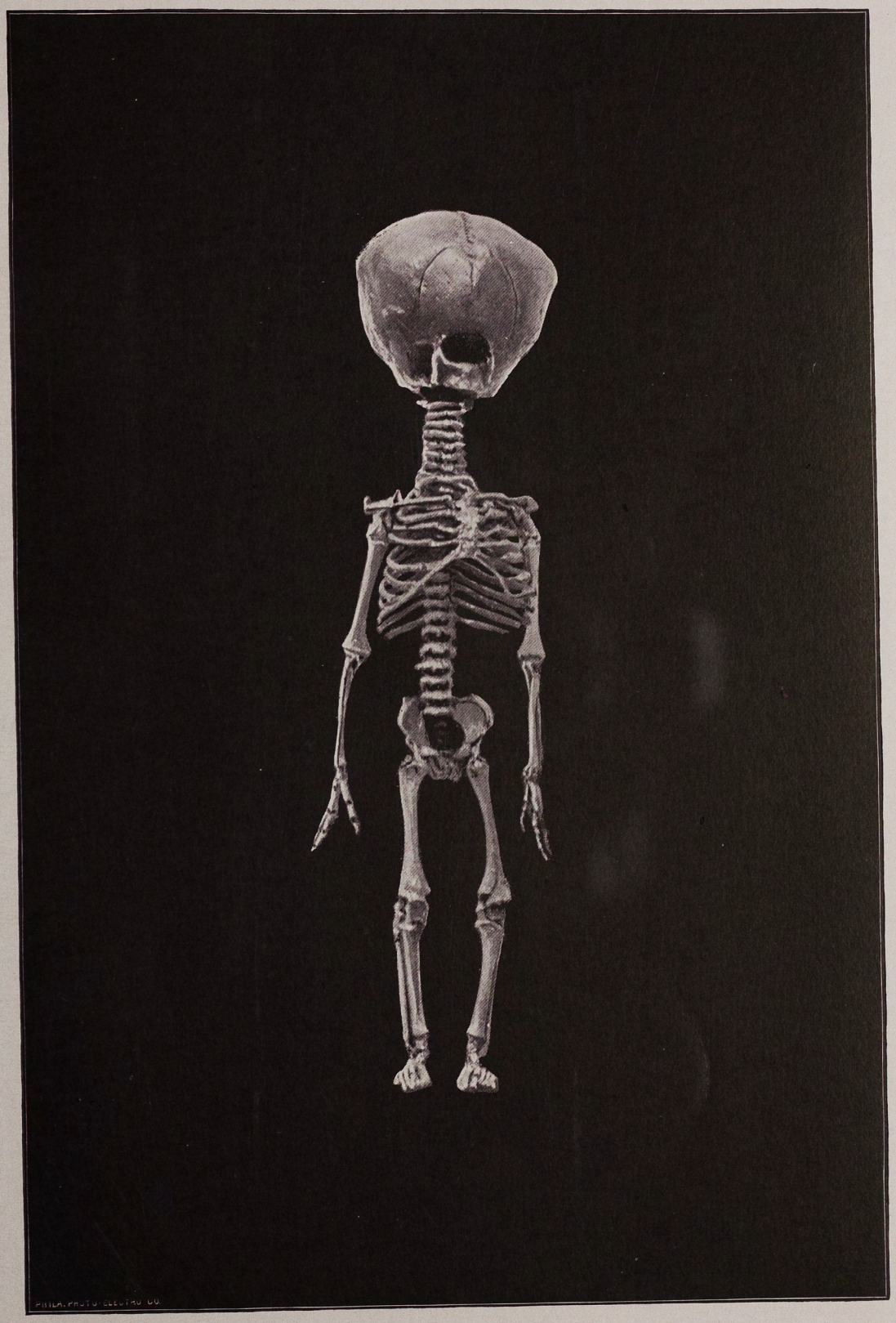
RHINOCEPHALUS.



CYCLOCEPHALUS.

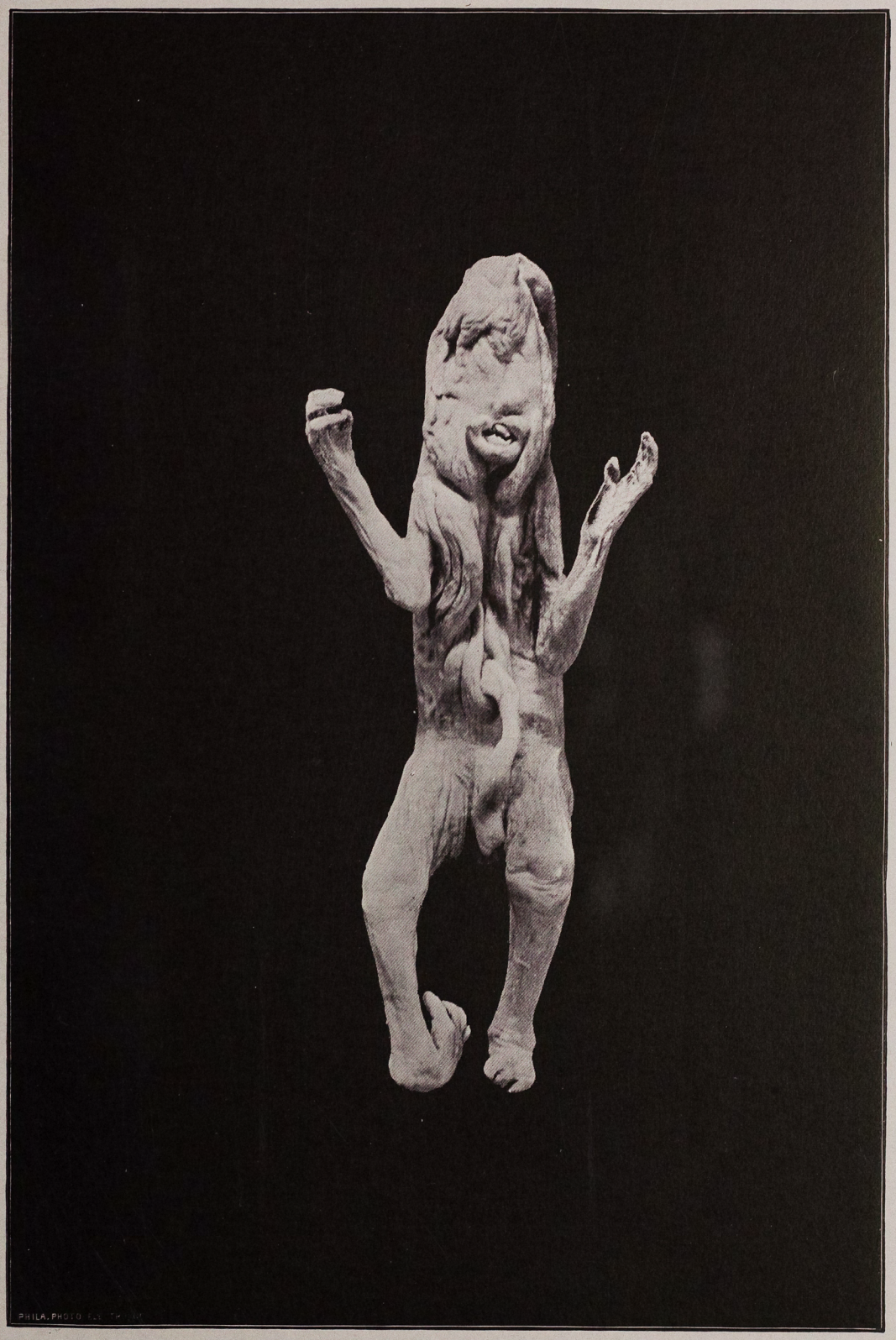
(CYCLOPS.)

PLATE XXIII.



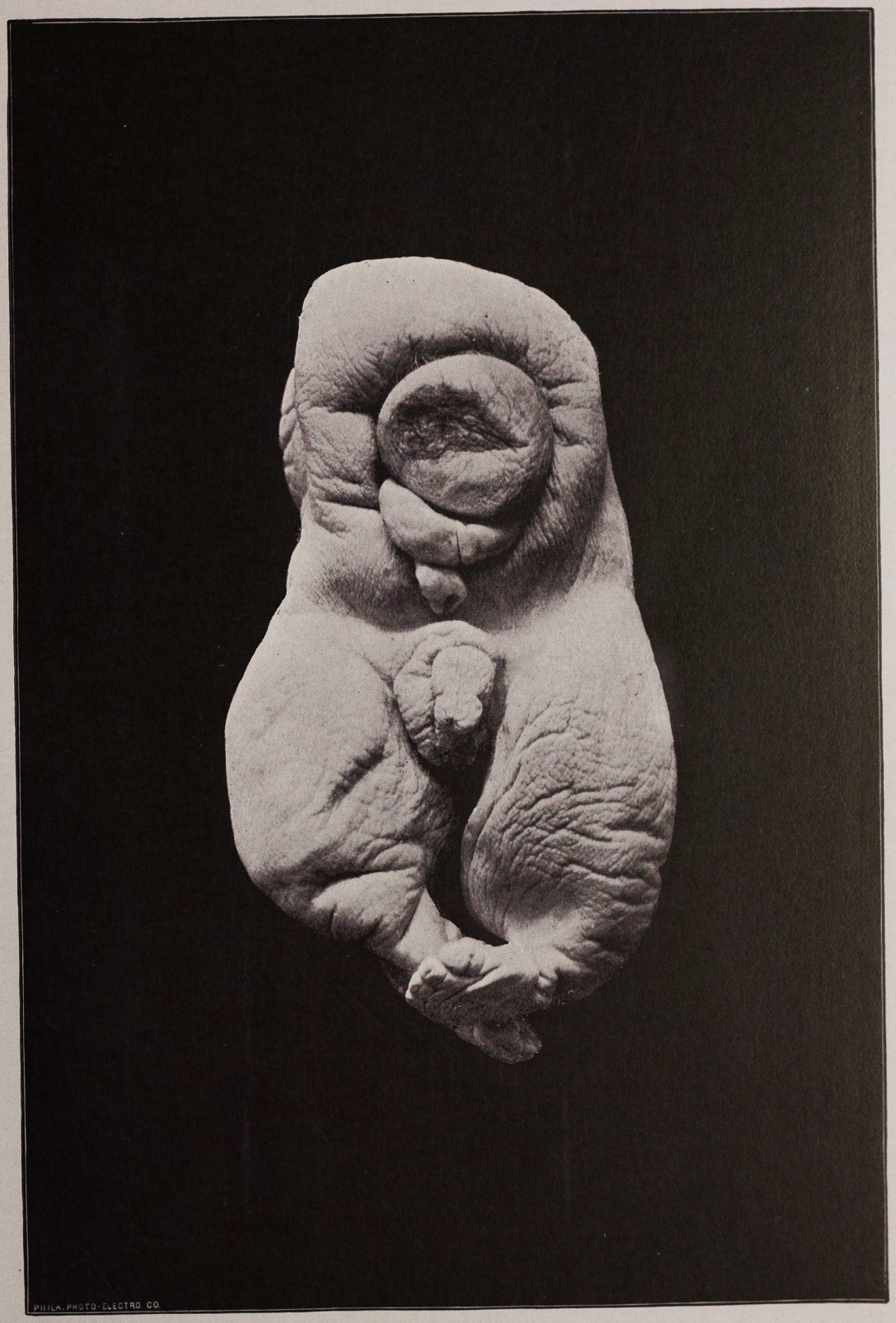
SKELETON OF CYCLOCEPHALUS.

PLATE XXIV.



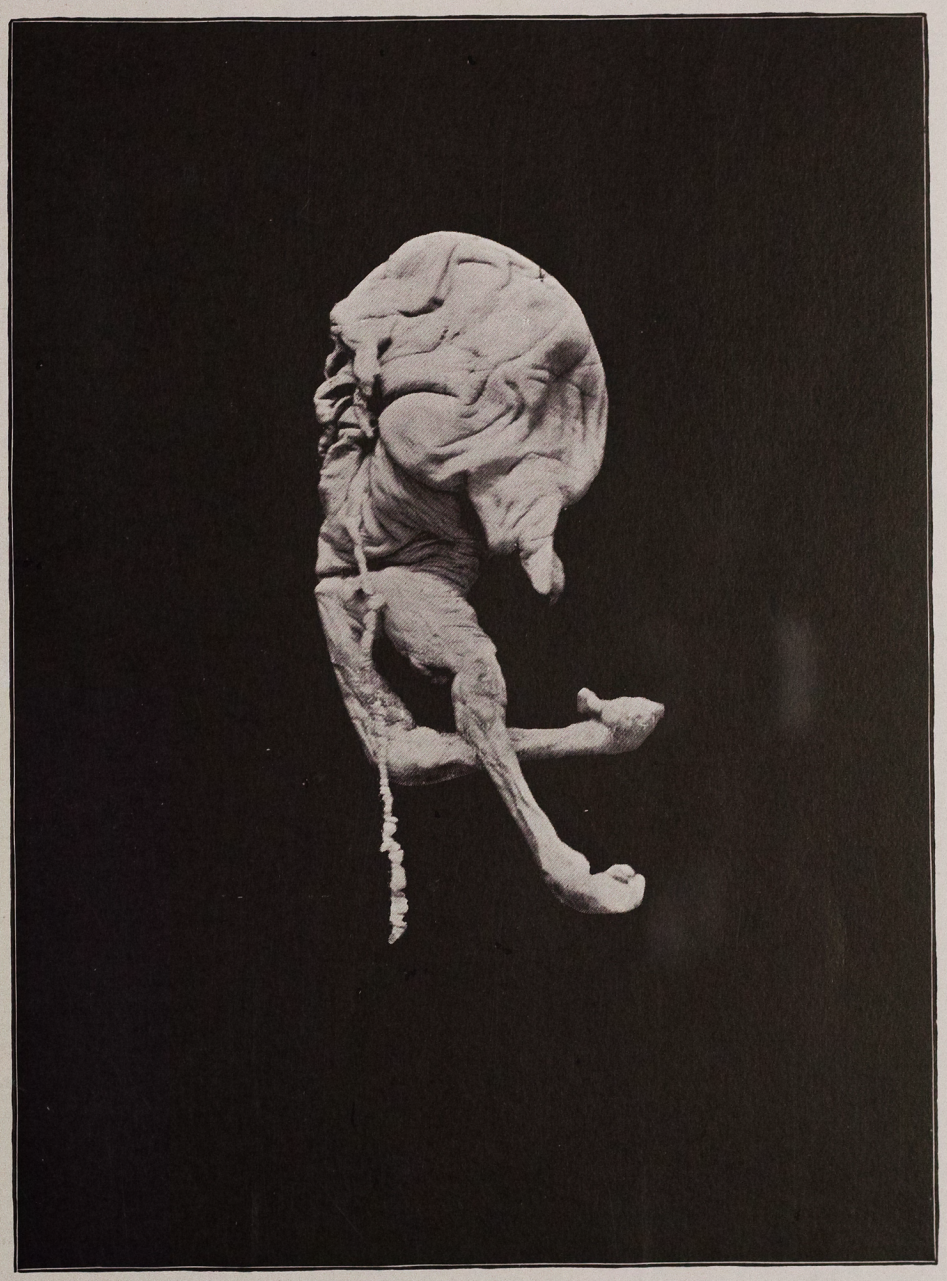
PARACEPHALUS.

PLATE XXV.



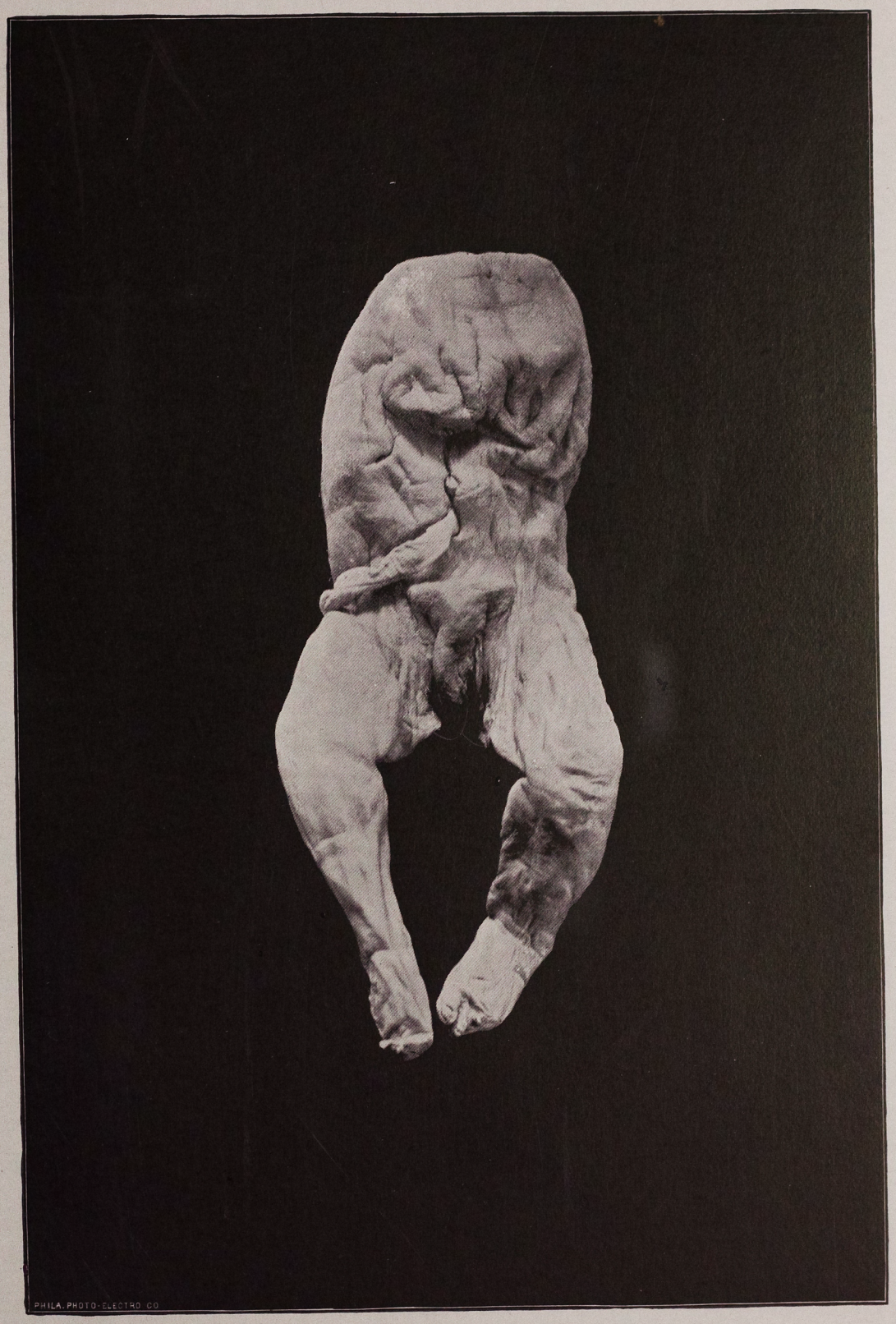
ACEPHALUS.

Plate XXVI.



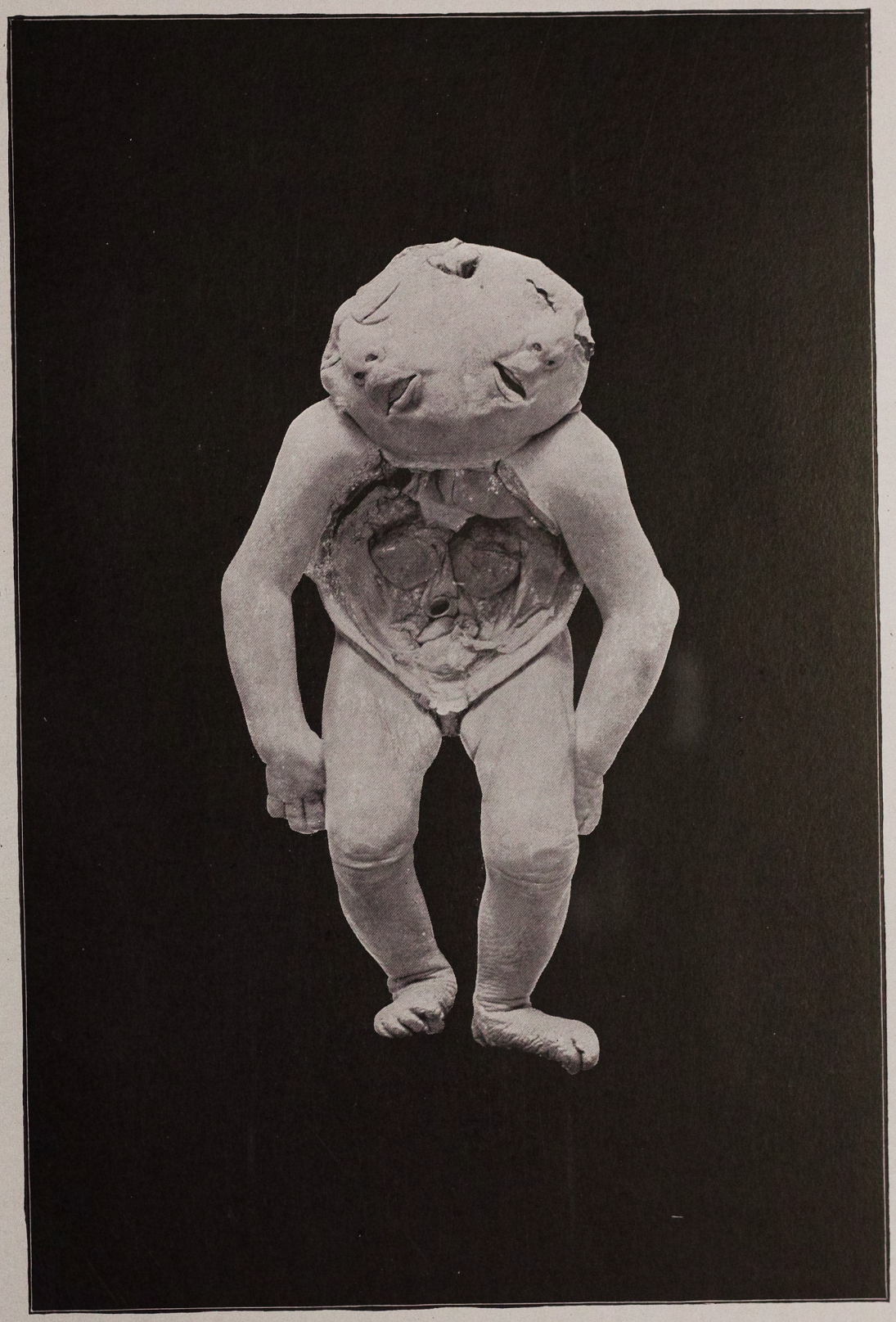
ACEPHALUS.

Plate XXVII.



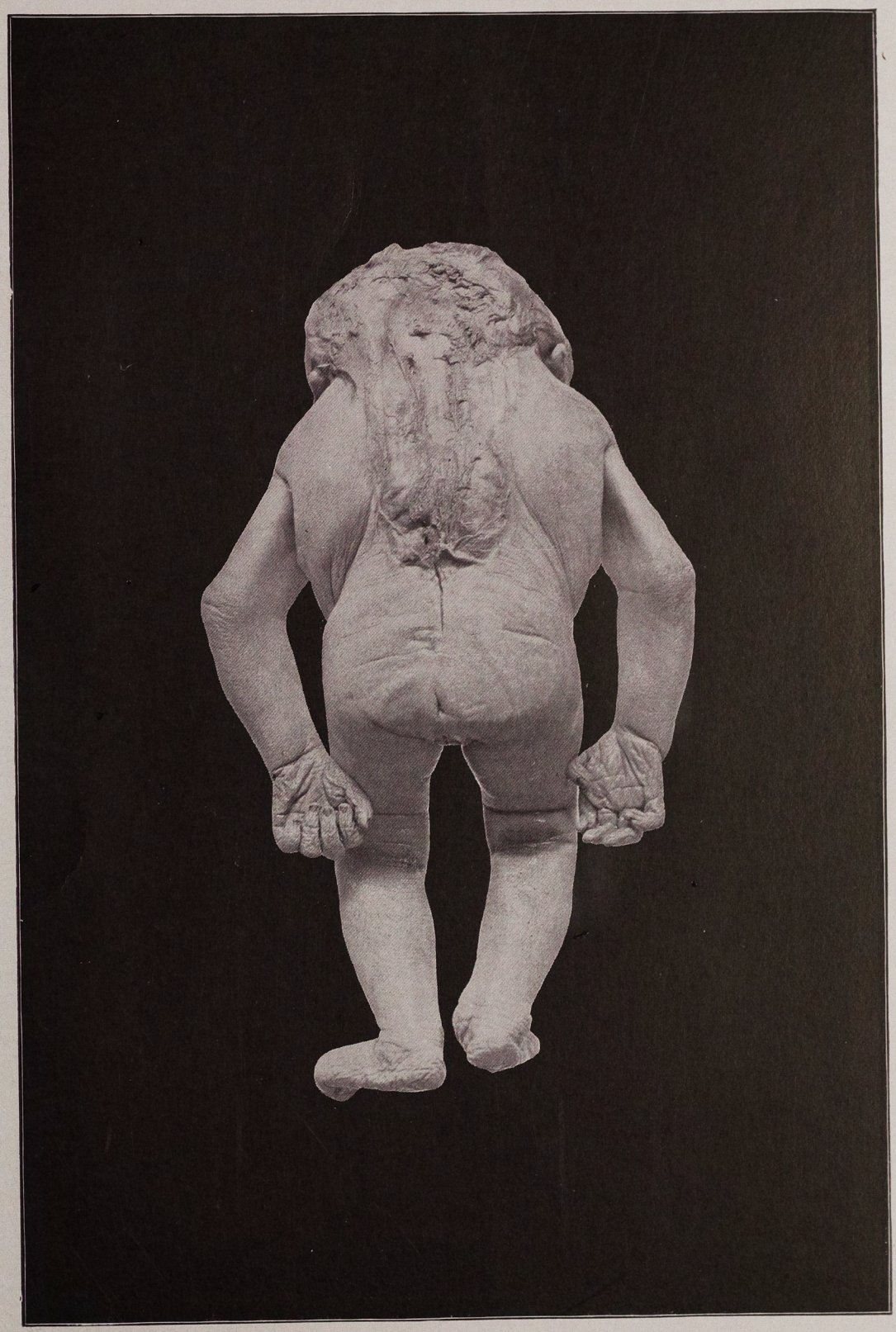
ACEPHALUS.

Plate XXVIII.



DIPROSOPUS.

PLATE XXIX.



DIPROSOPUS.

(REAR VIEW.)

No. .....

THE PROPERTY OF

THE HARTFORD MEDICAL SOCIETY,

HARTFORD, CONN.

Presented by Edward K. Root M.D. 1893.